



Adolescents with Tuberous Sclerosis Complex

Introduction

This fact sheet will provide an overview of the “normal” developmental tasks of adolescence while concentrating on the particular health care needs and challenges as faced by the teenager with Tuberous Sclerosis Complex (TSC). There are many excellent books on parenting a teenager as well as raising a disabled child. Please refer to additional Fact sheets that have outlined the specific issues related to the child and adult with TSC and concurrent attention difficulties, autism, learning disability and other psychiatric and behavioural difficulties.

Normal Adolescent Changes

Adolescent years are marked by changes in almost every aspect of the teenager’s life. Adolescents are developing a strong sense of their identity and discovering their personal values, adjusting to dramatic body changes, developing abstract thought, acquiring interpersonal skills, establishing independence from their family while negotiating a new relationship with their family, setting goals for future achievement and choosing a vocation. It is important to recognise some of the normal behaviours and reactions of parents and teenagers during this period. Many parents may perceive certain teen attitudes and behaviours to be problems. In reality those attitudes and behaviours may reflect very typical adolescent patterns totally aside from the problems of an adolescent with a chronic illness such as TSC.

What Is Normal?

Early adolescence begins with the teenager asking themselves questions “Am I normal?” “Will I develop okay?” Teenagers look to their parents and peers for solving these questions and for affirmation that they are okay with the world.

Mid-adolescence is characterised by the teenager asking more in-depth questions “Who am I?” “Am I accepted?” “Do I have power to make decisions about myself?” Peers become more important in helping to

answer those questions, while parents may have less of an influence.

In late adolescence the teenager may ask, “Am I smart enough?” “Am I attractive?” “What can I do with my life?” The peer group becomes extremely important in influencing behaviours, attitudes, dress and values.

For teenagers with a chronic life long illness the developmental problems normally associated with adolescence seem magnified. The overwhelming concerns relate to social acceptance. Peer acceptance is extremely important to a teenager and a teenager’s perception of his or her physical appearance often determines psychological adjustment. Adolescence is a time of significant changes. Achieving independence from family, formulating values and self-concept, and planning for the future become important objectives.

How moody should a child be? How talkative, rebellious, oppositional, or resistant? What is normal teenage behaviour? Why does my teenager appear to be so angry all the time? These questions are difficult to answer specifically. In general, normal behaviour is behaviour that does not interfere with a person’s ability to cope with his environment or to get along with others. But how do you determine what is a normal amount of moodiness in your teenager with TSC? Other Fact Sheets outline the specific issues that TSC may have in influencing the child with learning, hyperactive or impulsive behaviours, speech and communication. Many adults with TSC have reported feelings of anxiety, depression and anger. (See Fact Sheet 39 *Psychiatric and Behavioural Issues in Adults with Tuberous Sclerosis Complex*)

Some questions to ask yourself regarding your teenager’s behaviour may include:

1. How often does the behaviour occur?
2. Does the behaviour interfere with the teenager’s ability to function in the environment?
3. Does the behaviour interfere with others such as siblings or classmates, teachers, coaches, neighbours, and others whom they may have daily contacts with?
4. Have you considered your teenager’s individual differences? How different is the behaviour or attitude when compared with other children in their age group?

Special Problems In Adolescents With Tuberous Sclerosis

Epilepsy

An estimated 80 to 90% of all patients with TSC experience a first seizure with approximately 27% occurring from 1 to 16 years (Franz). The kind of seizures that the child with TSC has been experiencing in childhood may change as adolescence approaches. These changes may not indicate any significant problem but simply mean the child's nervous system is growing and developing into a mature adult's. Some adolescents may not experience a seizure until their teen years and may present with sudden or unusual alterations of their behaviour. It is very important to clarify the episodic event with careful observation and documentation. Complex partial seizures may present as unusual changes in behaviour and teenagers may experience a variety of "somatosensory" symptoms associated with these seizures. Complex partial seizures are those seizures that involve only part of the brain (usually the temporal or the frontal lobes) and those alter consciousness or awareness. It may be accompanied by automatisms. These are movements that often consist of smacking of the lips, chewing, picking at clothes, or wandering about in a confused fashion. Teenagers may have difficulty in finding the correct word to say or they may know the correct word yet be unable to say it, or say it clearly. They may have difficulty in comprehension of words or phrases. Teenagers experiencing complex partial seizures may describe a variety of unusual sensations of the abdomen (cramps or nausea) or body sensations such as a certain smell, usually an unpleasant, strong smell such as the smell of burning rubber. Some seizures may begin with the sensation of fear, followed by changes in blood pressure, heart rate and the colour of their skin (either paleness or facial flushing). Teenagers may describe flashbacks or strong memories or feelings as if something had been seen or experienced previously (*déjà vu*). Complex partial seizures may create or recreate one or all, or any combination of these feelings or experiences.

Some questions to ask yourself regarding these episodes may include:

1. What setting did the event occur?
2. What was the description of the event from beginning to end?
3. What happened first, second or third?
4. What kinds of movements were noticed? Was one side of the body affected or did the movements move to the other side as well?
5. How long did the event last? Were you able to time it?

6. During the episode did the teenager seem to have lost the ability to communicate with you or understand you?
7. After the episode, was the return to their normal behaviour immediate or seem delayed?
8. If the return to normal or "baseline" behaviour was delayed was the teenager able to speak or function normally?
9. Was the speech pattern normal?

Between 10% to 20% of all children in the United States are affected by chronic illness, mostly due to a genetic disorder. For the adolescent with a chronic disease such as TSC and epilepsy it can be more complex. The goal for our teenagers as they grow older will be for them to become more responsible for more of their own care, while teaching independence and responsibility for their health. Adolescence is the time when teenagers should begin the transition from childhood into independent adulthood. The transition is a gradual process beginning in childhood so that by adolescence they will have the skills in order to take care of their epilepsy and other health conditions associated with TSC. This may mean that they know the names of their medications they are taking, the doses, and the times of the administration. They should be responsible for keeping track of their seizure frequency if they have recurrent seizures. Teenaged patients should be fully informed about their epilepsy and should be allowed to make their own decisions, within the limits of reason and experience. Allowing a teenager to make a choice may mean the teenager has the right to make choices about medication and therapeutic recommendations. At some point the teenager may refuse or avoid taking their medication on a regular basis. This may be expected and should be dealt with as a matter of fact, emphasising the risk of seizures. All parents worry that something bad may happen to their child but allowing your child the opportunities to take risks and become responsible for their own health needs ultimately will help the adolescent to achieve good emotional and social development.

Teenagers with epilepsy and TSC must have education about their condition and advised about the possible implications of epilepsy and treatment on adult issues, such as birth control and family planning. Teenaged girls must be aware of the effect of anticonvulsants on the unborn baby as well as the genetic risk involved with future pregnancy. Genetic counselling should be encouraged for all adolescents affected with TSC. Most adolescents want to be given specific information about their reproductive potential, and particularly the probability of passing on TSC to the next generation. Anxiety regarding the uncertainty of these life long decisions will often surface. Adolescents with epilepsy

may be at greater risk for depression and suicidal ideation than their peers. The possibility of pregnancy in adolescence has major implications for the teenager with TSC. Young women should understand the interactions between anticonvulsants and contraceptive pills. Many anti-seizure medications change the effectiveness of birth control pills and these teenagers may need higher doses or stronger birth control. Young women with TSC must be informed about the potentially adverse effects of epilepsy and seizure medication on the developing foetus. It is always wise to consider questions about making any changes in medication or reducing medication before conceiving a baby. In other words, PLAN AHEAD. If there were a concern that the adolescent female would become pregnant, Folate 1mg daily is usually recommended for prevention of neural tube defects (spina bifida).

Both boys and girls need to understand the potentially adverse effects of alcohol and substance abuse on epilepsy and the potential to make their seizures worse. Seizures may increase in frequency or change during the rapid growth that occurs during adolescence. Seizure frequency, quality or duration of seizures may change as a reflection of changes in the size of subependymal nodules. These lesions rapidly grow between birth and 18 years of age. Girls may notice that their seizures increase prior to, or during, their menstrual cycles. Sleep, or lack thereof, is already a documented problem in those with TSC and may also interfere with seizure control. Promotion of good health habits, encouraging proper rest and exercise, as well as good nutrition, are essential to any plan for managing epilepsy during the busy and active schedule that teenagers keep.

Compliance with medical issues and recommended therapies in adolescents with epilepsy is often a difficult issue. A recent Finnish study of 300 adolescents with epilepsy ages 13-17 revealed that 22% of adolescents with epilepsy followed suggested health regimens, while 44% placed themselves in the category of 'satisfactory compliance', and the remaining 34% reported poor compliance. Compliance with their recommended life style was poorest, while the highest degree of compliance was recorded for medication. Compliance is especially important if the teenager is driving a motor vehicle. Almost all teenagers want to be able to drive an automobile. Sometimes their only motivation to be compliant with medication hinges upon obtaining a driver's license. It is often helpful to gently remind young teenagers that driving will not be possible until their seizures are controlled for a specific length of time and that there is assurance that medication is taken as prescribed on a regular basis.

Kidney

Because there are so many important changes to other body systems during adolescence the teenager should be educated regarding other manifestations of TSC and potential changes that would warrant seeking medical treatment. Other than central nervous system involvement the kidney is the next organ system most frequently affected. Renal cysts that cause symptoms often represent the earliest manifestation of TSC that have often developed in infancy. If there is known renal (kidney) involvement such as kidney cysts in association with TSC teenagers are monitored for the development of hypertension (high blood pressure) that often precedes the development of more functional impairment of the kidneys. Controlling high blood pressure is very important in the care of teenagers with severe cystic involvement of the kidneys as well as routine monitoring of kidney function by laboratory or diagnostic testing. Compliance with anti-hypertensive medications is very important.

AMLs (angiomyolipomas) are the most common kidney lesions in patients with TSC. AMLs appear somewhat later in life (young adulthood) and generally do not cause any symptoms, unless there would be sudden haemorrhage that may lead to a potentially life threatening situation. If the teenager should experience blood in the urine, dull pain in the flank area, lower back or abdominal area this may represent internal bleeding and may indicate a change that would need immediate medical attention. Teenagers with TSC should have their kidneys imaged on a regular basis every 1-2 years. The renal lesions may remain stable for long periods of time and may not require any specific treatment. Rarely, renal cell carcinoma or cancer in the kidney is seen. The tumours should be removed as soon as possible.

Skin

Skin manifestations are an important part of TSC and are reported to occur in more than 80% of children over 5 years of age. Because body image is so important to a developing teenager, sensitivity to addressing their concerns regarding these facial features and appropriate treatments is critical. As a child enters puberty, the typical skin lesions may become even more apparent. The facial angiofibromas can be removed using dermabrasion or other laser techniques. Research has suggested that the facial angiofibromas should be removed using laser treatment when they are small and less fibrous.

Letting Go

Of all the challenges facing the parent of a child with a disability or chronic illness, it is both essential and the most difficult to encourage independence and

make the transition to adulthood. While the grief of a parent or sense of parental responsibility cannot be underscored it is vital that you encourage, support, and help motivate your child/teenager's independence. Your goal should be to make your role much less important as years pass. Problem solving together, while nurturing independence and treating or thinking of your teenager "as if" they are capable of achieving their potential for living and working, is important. Helping your teenager to learn more about their disease and how to manage the many aspects of it are crucial for understanding and living with TSC as well as accepting it. Parents of children who have a problem of any type are prone to overprotect the child. If your teenager has epilepsy, the problem is often worse because of the unpredictable nature of seizures. Parents of all teenagers must learn to gradually allow the teenager to take more risks as they develop. Overprotecting your teenager should be avoided as it may have more serious impact on emotional and social development.

Accepting the Diagnosis

Teenagers will feel different, and isolated, and may experience poor self-image as angiomas become more apparent or as flares of seizures occur. The teenager at some point will have to confront their own disability or health problem while adjusting to the changing demands of these difficult years. Teenagers may act out at school or withdraw. Compliance with medication or following suggested routines may be a struggle for a teenager confronting their health problems or disability. Encouraging social relationships help in preventing isolation and loneliness. Talking with other teenagers with similar issues, or seeking professional counselling, is often necessary to help

teenagers to change their self-image and boost their confidence. Issues of planning for the future and what to expect in adulthood may seem extremely overwhelming to a teenager unsure of themselves and of their diagnosis. Help your teenager to build support systems outside of the family. As the adolescent discusses their illness with their friends it allows them to develop positive life skills necessary to manage their care (i.e. keeping scheduled appointments, when to take medicine or seek the advice of their physician, moderate physical activities). Encourage the adoption of behaviours that are favourable to their health and reinforce these behaviours (participating in extra-curricular activities, jobs).

Conclusion

Raising a teenager can be a very frustrating experience. However, understanding the unique perspective that your teenager may have is a first step in helping your teenager with the transition to adulthood and independence. Epilepsy can make parenting more difficult. TSC presents unique problems in parenting and difficult challenges. Studies involving chronically ill adolescents have identified how parents may need to remain very involved in their adolescent's health care but at the same time encourage growth and independence into adulthood.

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Tuberous Sclerosis Association



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